CASE REPORT

A case report of a right ventricular mass in a patient with Behçet's disease: Myxoma or thrombus?

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Introduction: **Behçet's disease (BD) is a multi-system, chronic and relapsing disorder classified as ''vasculitic syndrome''. It typically affects young adult females between 20 and 40 years of age. There are some typical clinical manifestations associated with this disease, however, at times; rare sign and symptoms pose a challenge to the treating physician and making a definitive diagnosis. Presentations with cardiac symptoms are one of the extremely rare manifestations of the Behçet's disease.**

Methods: **The authors present clinical, laboratory and imaging findings of a patient who presented with a cardiac mass which was the first presenting feature or manifestation of Behçet's disease.**

Results: **A 19-year-old boy was admitted to our hospital for the investigation of ''fever of unknown origin'', weight loss, shortness of breath and a scrotal ulcer of recent on-set. X-ray chest and electrocardiograms were inconclusive. Transthoracic echocardiography revealed a right ventricular (RV) mass attached to the interventricular septum measuring 1.5 × 1.5 cms (Panel A). Cardiac MRI identified it as a RV Myxoma. In addition, on CT scan of the chest pulmonary embolism was noted. The patient underwent excision biopsy of the tumor under cardiopulmonary bypass via right atriotomy (Panel B). Histopathology of the mass described it as ''an organizing thrombus with a few groups of interrupted myocardial fibers and some infiltration of lymphocytes and plasma cells''. Moreover his HLA typing was found positive for HLA-B51 (5). In view of the above findings and associated lesions, the patient was diagnosed as a case of Behçet's disease. The medical management included immunosuppressant and anticoagulation.**

Conclusion: **Behçet's disease, even in the absence of the typical clinical features, should be considered in the differential diagnosis of right ventricular mass, especially when dealing with young adults from the Mediterranean basin and the Middle-East.**

Keywords: Right ventricular mass, Scrotal ulcer, Behçet's disease, HLA-B, B51(5)

Introduction

Behçet's disease (BD) is a multisystem, chronic, relapsing condition classified as an auto-inflammatory syndrome, sharing some common immune and genetic mechanisms [1,2]. Despite its common manifestations, presentations with cardiac symptoms are one of the rare manifestations of BD, posing a challenge for the treating physician. During the current era of highly

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sophisticated cardiac imaging techniques such as Echocardiography, Computed Tomography Scan and Magnetic Resonant Imaging (MRI), more and more reports on cardiovascular involvement, predominantly related to right-sided heart, have been published [3–11]. The authors are reporting this case to highlight this relatively rare presentation of BD which might be of interest for physicians and surgeons.

Case history

A 19-year-old Jordanian male was referred to an out-unit for the surgical removal of an RV mass. The gentleman had a history of weight loss and fever over two months and a remote past history of recurrent oral ulcers. On physical examination, the patient had a fever of 37.8 °C, a right scrotal ulcer and multiple scattered indurations at the previous venipuncture points. The systemic examination was otherwise unremarkable.

A repeat echocardiographic examination confirmed the presence of a 2.1 × 1.5 cm, rounded mass of non-uniform echogenicity attached to the right side of IVS (Fig. 1). A Doppler ultrasonographic scan of both lower limbs was negative for deep vein thrombosis and a pulmonary ventilation/perfusion scan showed a mismatched, peripheral wedge-shaped perfusion abnormality involving part of the superior segment of the right lower lobe suggesting sub-segmental pulmonary infarction. Cardiac MRI reported it as a well-defined, round shape, smooth, inhomogeneous and pedunculated mass, arising from the interventricular septum and protruding into the apical aspect of the RV. There was partially increased signal intensity on the T2 weighted sequence and moderate enhancement following gadolinium, the features favoring the diagnosis of a RV myxoma. In context with the history of fever, the possibilities of an RV myxoma, infected vegetation or an intracardiac metastatic lesion were considered. Owing to the presence of fever, scrotal ulcer and indurations at the previous venipuncture points (Perthes' response) and a past history of recurrent oral ulcers, BD was also thought as one of the primary diagnoses and the RV mass could be a thrombus due to BD.

Considering the potential risk of mechanical obstruction of the pulmonary valve by the mass, its embolization to the pulmonary circulation with fatal consequences, as well as, to have a histopathological diagnosis, the patient underwent surgical excision of the tumour. Macroscopically, it was found to be an oval shaped mass, measuring 1.2 × 0.7 × 0.3 cm at its greatest dimension (Fig. 2).

Figure 1. Echocardiographic view of the mass in right ventricle.

Figure 2. Macroscopic view of the excised mass.

Histopathological examination revealed an organizing thrombus with a few groups of interrupted myocardial fibers and some infiltration of lymphocytes and plasma cells (Fig. 3). Serological profile was negative for antinuclear antibodies and rheumatoid factor. Three sets of blood culture did not show any growth. Human leukocyte antigen analysis detected HLA-B, B51 (5). Aforementioned data collectively confirmed the diagnosis of BD and RV thrombus as one of its rare presentations. The patient was put on heparin, warfarin, corticosteroids and azathioprine and discharged from hospital a week after the surgery. Follow up TTE was cleared of RV mass (Fig. 4).

Discussion

BD is a chronic inflammatory disorder with multisystemic manifestations. Cardiac involvement is a rare occurrence and has ranged from 7% to 29% of reported cases [12]. In one series of analy-

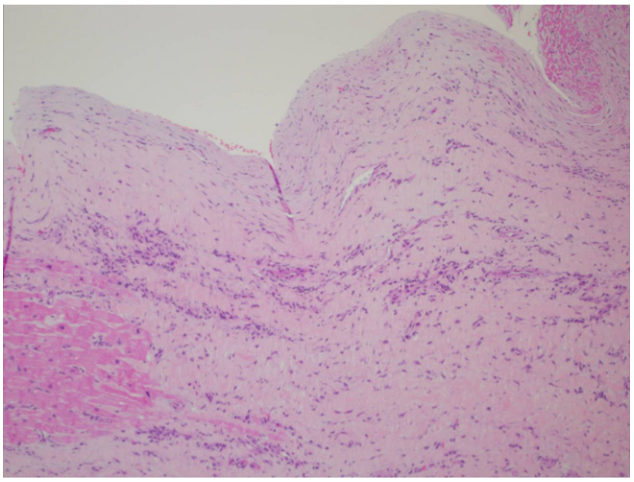

Figure 3. Histopathological slide of the mass.

sis of 2147 patients, the author found only three patients with cardiac involvement [13].

BD has high prevalence in Japan and eastern Mediterranean countries and is relatively low in United States and Northern Europe. It is most common in Turkey (80–370 cases per 100,000) [14]. It typically affects young adults between 20 and 40 years of age and females are outnumbered by males.

To date, no specific etiological agent responsible for this disorder has been identified. Clinical, immunological and genetic features of BD led to its classification within the frames of systemic vasculitides, seronegative spondyloarthritides and, more recently, auto-inflammatory disorders [1,15].

Conventionally, the syndrome is diagnosed by the symptoms of recurrent oral ulcers, genital ulcers, uveitis, arthritis, skin lesions and nervous system involvement. The Behçet's Disease Committee of Japan and International Study Group for Behçet's Disease have proposed major and minor criteria [16,17]. Both, however, lack the appreciation of cardiac involvement in BD which might manifest as pericarditis, cardiomyopathy, myocarditis, endocarditis, valvular dysfunction, endomyocardial fibrosis, conduction abnormalities, coronary artery disease, pseudo-aneurysm, rupture of the sinus of Valsalva and intracardiac thrombus [18–20].

The exact pathological mechanism of cardiac thrombus formation in BD is unknown. In most cases, the histological description of the thrombus and underlying myocardium suggests an inflammatory process with dominantly mononuclear cell infiltrate. On the other hand, in some cases, biopsy findings included endomyocardial fibrosis or normal myocardium. A high level of erythrocyte sedimentation rate, C-reactive proteins, antiphospholipid antibodies, anti-endothelial antibodies, von Willebrand factor and plasma endothelin-I level support an auto-immune and inflammatory theory.

In BD, the site of the intracardiac thrombus is variable. In an analysis of 25 patients, more than half were found to have thrombus in RV (52%). RA was the second most commonly involved cavity (24%). Left atrium and left ventricle were the least affected chambers (4% each) and multichamber thrombi were found in 16% of the subjects [21]. It is unclear why the thrombi in BD have an increased propensity for right-sided chambers.

Although surgical exploration and autopsy findings suggest that the thrombus is tightly attached to the underlying myocardium, it is still considered a potential source of distal embolization, particularly to the lungs resulting in pulmonary embolism and infarction as was found on the pulmonary ventilation/perfusion scan in our patient. Moreover, the physical presence of the thrombus may cause valvular and atrioventricular dysfunction, resulting in symptoms of right-sided heart failure.

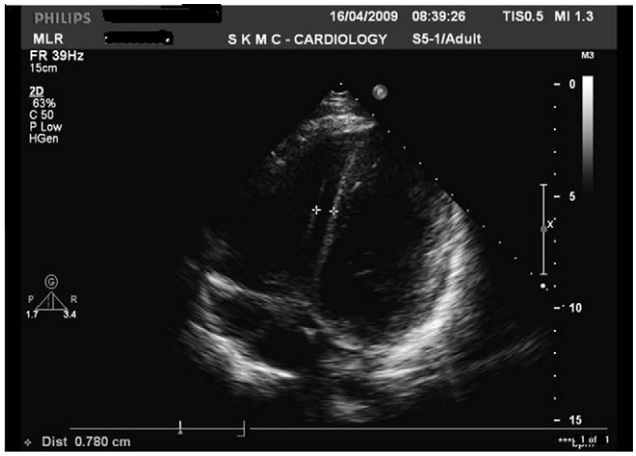

Figure 4. Echocardiographic apical four chamber view after the excision of the mass from the right ventricule.

BD involving the heart in general has very poor prognosis and mortality can be as high as 20% in the months or years following diagnosis [22]. The primary goal of management is immunosuppression and immunomodulation. Uncertainties surround the issue of anticoagulation as no controlled data exists on the benefits of anticoagulants in the management of cardiac thrombi in BD. Recently released guidelines for the management of BD [23] do not discuss the use of warfarin, specifically in reference to intracardiac thrombus in this condition. Nevertheless, a substantial number of case reports have been published wherein vitamin-K antagonist has been effectively used [24,5,25]. Our patient was treated successfully with azathioprine, steroids, and warfarin. Reservations about the use of anticoagulation in BD remain [26] and further studies are warranted to provide the clinician with recommendations over the safety and effectiveness of anticoagulation in cardiac BD. Cardiac surgeons may be involved in certain potentially life-threatening associations such as in the index case.

Conclusion

The authors believe that in the presence of constitutional symptoms, BD should be considered in the differential diagnosis of the right ventricular mass, especially when dealing with young adults from the Mediterranean basin and the Middle East. The initial diagnosis of cardiovascular BD can be extremely difficult, especially in the absence of its florid clinical manifestations. On occasion, cardiac symptoms may be the presenting complaints and may precede other signs and symptoms essential for the diagnosis of this disease by many years. Thus upon finding an intracardiac mass, with a history of weight loss, fever and other non-specific symptoms, BD should be considered as one of the possibilities in addition to more commonly known conditions like myxoma and vegetation. Familiarity with this rare cardiac condition might help in early diagnosis and treatment of BD.

Competing interest

None.

Authors' contributions

All authors read and approved the final manuscript.

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